

# Neutrophilic Dermatitis of the Dorsal Hand: A Distinctive Variant of Sweet Syndrome

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## PRACTICE POINTS

- Neutrophilic dermatosis of the dorsal hand (NDDH) is a reactive neutrophilic dermatosis that includes Sweet syndrome (SS) and pyoderma gangrenosum.
- Localization to the dorsal aspect of the hand, presence of ulcerative nodules, and older age at onset are characteristic features of NDDH.
- Meeting the major criteria alone for SS may be a more sensitive way to diagnose NDDH, as serum inflammatory markers may not be remarkably elevated in this condition.

To the Editor:

Neutrophilic dermatosis of the dorsal hand (NDDH) is an uncommon reactive neutrophilic dermatosis that presents as a painful, enlarging, ulcerative nodule. It often is misdiagnosed and initially treated as an infection. Similar to other neutrophilic dermatoses, it is associated with underlying infections, inflammatory conditions, and malignancies. Neutrophilic dermatosis of the dorsal hand is considered a subset of Sweet syndrome (SS); we highlight similarities and differences between NDDH and SS, reporting the case of a 66-year-old man without systemic symptoms who developed NDDH on the right hand.

A 66-year-old man presented with a progressively enlarging, painful, ulcerative, 2-cm nodule on the right hand following mechanical trauma 2 weeks prior (Figure 1). He was afebrile with no remarkable medical history. Laboratory evaluation revealed an erythrocyte

sedimentation rate (ESR) of 20 mm/h (reference range, 0-10 mm/h) and C-reactive protein (CRP) level of 3.52 mg/dL (reference range, 0-0.5 mg/dL) without leukocytosis; both were not remarkably elevated when adjusted for age.<sup>1,2</sup> The clinical differential diagnosis was broad and included pyoderma with evolving cellulitis, neutrophilic dermatosis, atypical mycobacterial infection, subcutaneous or deep fungal infection, squamous cell carcinoma, cutaneous lymphoma, and metastasis. Due to the rapid development of the lesion, initial treatment focused on a bacterial infection, but there was no improvement on antibiotics and wound cultures were negative. The ulcerative nodule was biopsied, and histopathology demonstrated abundant neutrophilic inflammation, endothelial swelling, and leukocytoclasia without microorganisms



**FIGURE 1.** Ulcerating nodule on the dorsal aspect of the right hand with surrounding inflammation.

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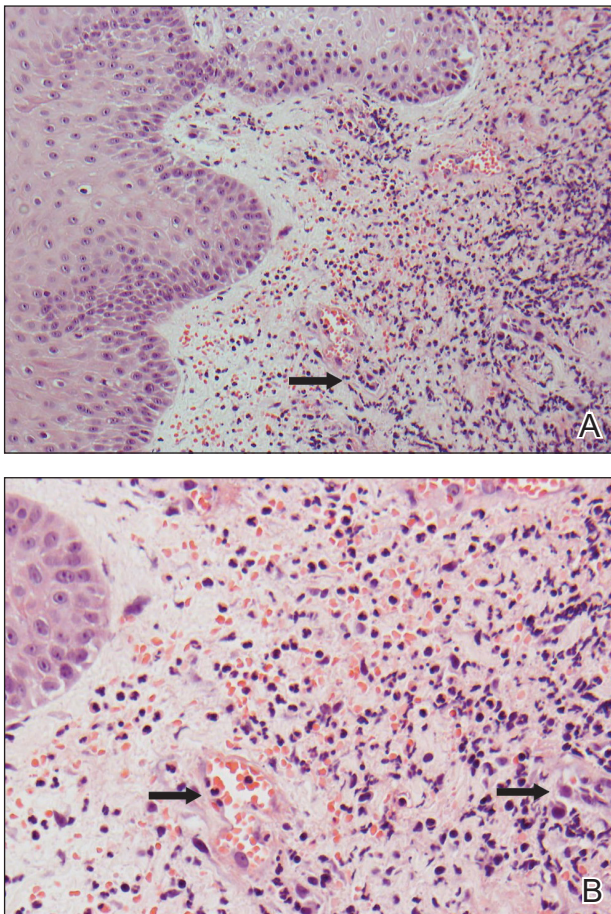
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(Figure 2). Tissue cultures for bacteria, fungi, and atypical mycobacteria were negative. A diagnosis of NDDH was made based on clinical and histologic findings. The wound improved with a 3-week course of oral prednisone.

Neutrophilic dermatosis of the dorsal hand is a subset of reactive neutrophilic dermatoses, which includes SS (acute febrile neutrophilic dermatosis) and pyoderma gangrenosum. It is described as a localized variant of SS, with similar associated underlying inflammatory, neoplastic conditions and laboratory findings.<sup>3</sup> However, NDDH has characteristic features that differ from classic SS. Neutrophilic dermatosis of the dorsal hand typically presents as painful papules, pustules, or ulcers that progress to become larger ulcers, plaques, and nodules. The clinical appearance may more closely resemble pyoderma gangrenosum or atypical SS, with ulceration frequently present. Pathergy also may be demonstrated in NDDH,



**FIGURE 2.** A, Histopathology showed neutrophilic inflammation around smaller capillary-sized vessels (H&E, original magnification  $\times 200$ ). A central small vessel (arrow) was completely obliterated by neutrophils and rimmed by leukocytoclastic debris. B, Endotheliitis (arrows) was seen, as evidenced by swelling and neutrophilic infiltration around the small capillary-sized vessels (H&E, original magnification  $\times 400$ ). The nuclei showed reactive changes of prominent nuclei, enlargement, and retained nuclear:cytoplasmic ratios. The background had extravasated erythrocytes, neutrophils, and lymphocytes.

similar to our patient. The average age of presentation for NDDH is 60 years, which is older than the average age for SS or pyoderma gangrenosum.<sup>3</sup> Similar to other neutrophilic dermatoses, NDDH responds well to oral steroids or steroid-sparing immunosuppressants such as dapsone, colchicine, azathioprine, or tetracycline antibiotics.<sup>4</sup>

The criteria for SS are well established<sup>5,6</sup> and may be used for the diagnosis of NDDH, taking into account the localization of lesions to the dorsal aspect of the hands. The diagnostic criteria for SS include fulfillment of both major and at least 2 of 4 minor criteria. The 2 major criteria include rapid presentation of skin lesions and neutrophilic dermal infiltrate on biopsy. Minor criteria are defined as the following: (1) preceding nonspecific respiratory or gastrointestinal tract infection, inflammatory conditions, underlying malignancy, or pregnancy; (2) fever; (3) excellent response to steroids; and (4) 3 of the 4 of the following laboratory abnormalities: elevated CRP, ESR, leukocytosis, or left shift in complete blood cell count. Our patient met both major criteria and only 1 minor criterion—excellent response to systemic corticosteroids. Nofal et al<sup>7</sup> advocated for revised diagnostic criteria for SS, with one suggestion utilizing only the 2 major criteria being necessary for diagnosis. Given that serum inflammatory markers may not be as elevated in NDDH compared to SS,<sup>3,7,8</sup> meeting the major criteria alone may be a better way to diagnose NDDH, as in our patient.

Our patient presented with an expanding ulcerating nodule on the hand that elicited a wide list of differential diagnoses to include infections and neoplasms. Rapid development, localization to the dorsal aspect of the hand, and treatment resistance to antibiotics may help the clinician consider a diagnosis of NDDH, which should be confirmed by a biopsy. Similar to other neutrophilic dermatoses, an underlying malignancy or inflammatory condition should be sought out. Neutrophilic dermatosis of the dorsal hand responds well to systemic steroids, though recurrences may occur.

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